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CASE REPORT

Craniofacial/Pediatric

Juvenile Xanthogranuloma on the Upper Lip

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Summary: We treated a patient with juvenile xanthogranuloma on the upper lip. A yellow, elastic, hard tumor on the upper lip was evident from birth, which gradually increased in size. The patient was examined at our department at the age of 7 months, at which time the mass extended from the upper lip to the nasal cavity and measured approximately 1 cm. There was a risk that the mass might obstruct the nasal cavity, and an incisional biopsy was conducted to obtain a definitive diagnosis. In histopathological testing, the patient was diagnosed with a juvenile xanthogranuloma. Part of the mass still remains on the upper lip, but has not increased in size during postoperative monitoring. Juvenile xanthogranuloma on the upper lip is extremely rare, and to the best of our knowledge, this is only the fourth case to be reported in the plastic surgery literature in English. In most cases, juvenile xanthogranuloma regresses spontaneously, and unnecessary surgery is to be avoided. The possibility of juvenile xanthogranuloma should always be considered for masses that increase in size in infants and young children, and it is important to reach a definitive diagnosis by skin biopsy. *(Plast Reconstr Surg Glob Open 2021;9:e3712; doi: 10.1097/GOX.0000000000003712; Published online 19 July 2021.)*

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a benign disease of unknown etiology that appears as a yellow mass on the skin, either from birth or during early childhood. It usually appears on the face, head, neck, or trunk, and is classified as single or multiple. In very rare cases, it may be associated with a systemic comorbidity such as chronic myeloid leukemia or von Recklinghausen's disease.[1] We treated a patient with a JXG extending from the upper lip to the right nasal cavity that was present at birth.

CASE HISTORY

A 7-month-old girl was born with a yellow mass growing from the right upper lip to the nasal cavity. She was examined at a local pediatric clinic, where JXG was suspected and a policy of watchful waiting was adopted. However, in June 2020, she was referred to our department because the mass had recently increased in size and was growing into the right nasal cavity. Her medical history was unremarkable, and she was born by normal delivery. On presentation, a clearly demarcated elastic hard yellow mass approximately 1 cm in size extending from the upper lip to the right nasal cavity was seen. The mass extended into the base of the right nasal cavity and obstructed the nasal cavity (Fig. 1). JXG was suspected as the most likely diagnosis from the clinical course and shape of the mass. Histopathological investigation was required to reach a definitive diagnosis. However, because the mass was located on the upper lip, where it was extremely obvious, an excisional biopsy could not be performed for aesthetic reasons. Moreover, if it was actually JXG, it could also regress spontaneously and we therefore decided to avoid unnecessary resection. Hence, we planned to conduct an incisional biopsy that would not deform the upper lip, with the goal of debulking the mass by tissue biopsy. This was conducted under general anesthesia in July 2020. A vertical skin incision was made to remove as much of the mass as possible (Fig. 2). The mass was clearly demarcated and was removed by cutting into a part of it. The base of the mass was on the orbicularis oris muscle, part of which was also resected. A simple wound closure was performed. Histopathological testing revealed the thinning of the epidermis and round to short spindle-shaped cell clusters in all layers of the dermis. Foamy mononuclear histiocytes and Touton giant cells were present in places, and JXG was diagnosed (Fig. 3). The patient's postoperative course was uneventful, and 9 months postoperatively, although part of the mass remained on the upper lip, it had not regrown. The upper lip was also well shaped, with satisfactory aesthetic outcomes (Fig. 4).

DISCUSSION

JXG was described by Adamson in 1905 as one of the non-Langerhans cell group of histiocytic proliferative disorders, and in most reported cases, it is described as

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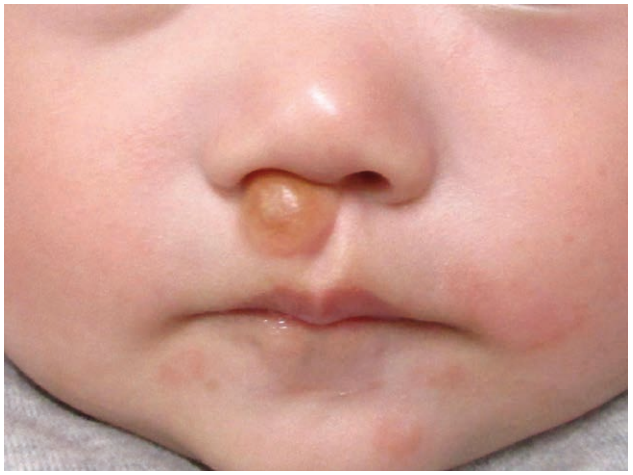

Fig. 1. Photograph on presentation. A clearly demarcated yellow mass extends from the upper lip to the nasal cavity.

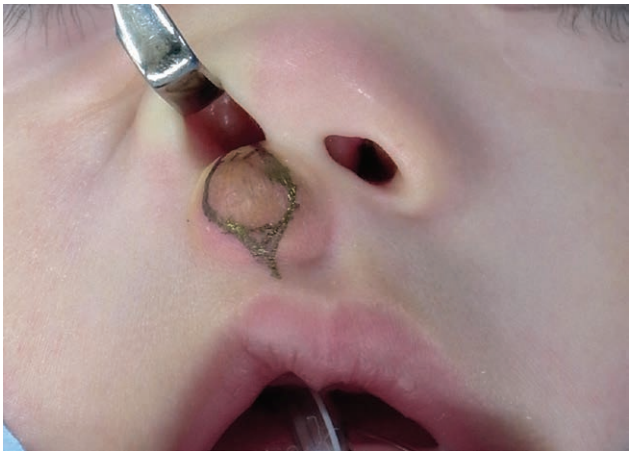

Fig. 2. Preoperative photograph. Vertical skin incisions were made to remove as much of the mass as possible.

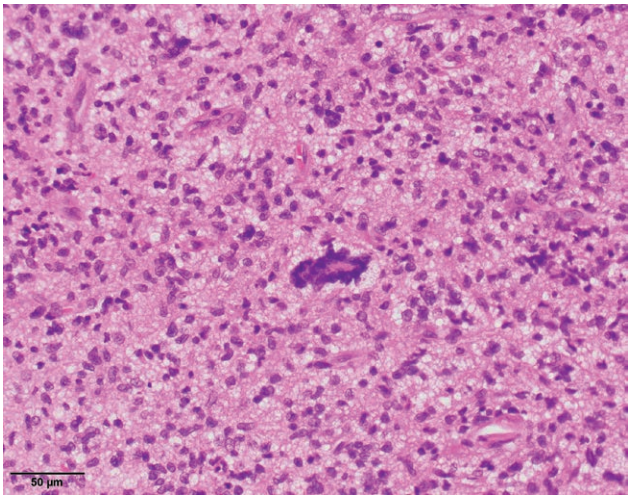

Fig. 3. Histopathological photograph (Hematoxylin Eosin staining). Histologically, no abnormality was evident, but foamy mononuclear histiocytes and Touton giant cells were present, and juvenile xanthogranuloma was diagnosed.

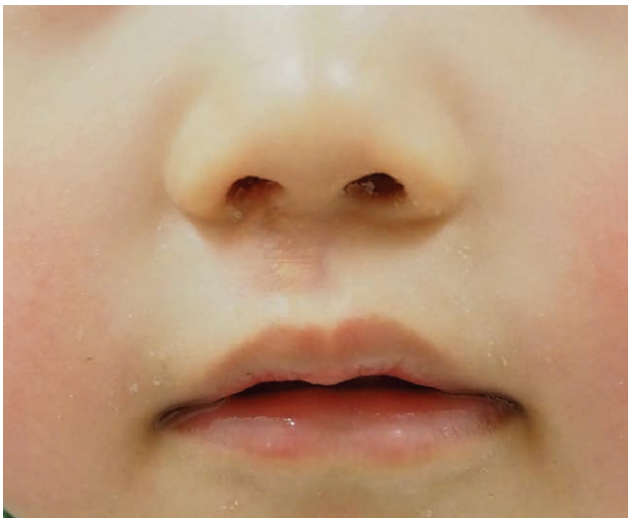

Fig. 4. A photograph at 9 months postoperatively. Although some parts of the tumor are still present on the upper lip, the tumor has not regrown. The upper lip and nasal cavity are well shaped.

a clearly demarcated, elastic hard, yellow skin nodule.[2] It occurs solely in 60%–82% of cases.[1] Gianotti and Caputo classified the clinical presentation of JXG into two types: the large nodular form, comprising one or a few nodules measuring 1–2 cm and the small nodular form, comprising multiple nodules measuring 2–5 mm; large nodules that exceed 2 cm in size are also referred to as "giant JXG."[3] Most cases occur within the first year of life, and although they grow rapidly, most spontaneously decrease in size within 3–6 years.[4] From the perspectives of both surgical invasion and postoperative scarring, watchful waiting is therefore the best policy in infants and young children. However, the mechanism of spontaneous regression is not well understood, and most patients with single JXG undergo resection with the additional purpose of definitive diagnosis.[5–7] The differential diagnosis includes dermatofibroma, xanthoma, and dermatofibrosarcoma protruberans.

In our patient, clinical presentation and other factors were strongly suggestive of JXG, although because the possibility of a different type of tumor could not be completely excluded, tissue biopsy was performed to obtain a definitive diagnosis. Because the mass was located in the middle of the upper lip, it threatened to obstruct the nasal cavity and reducing its volume was also urgently necessary from an aesthetic viewpoint. The mass also stretched the skin of the upper lip, which had become very thin, and even if it regressed spontaneously, it was very likely to leave an ugly scar behind. Therefore, it was decided to obtain a definitive diagnosis while preserving the shape of the nasal cavity and upper lip. As mentioned above, a high proportion of JXGs regress spontaneously within 3–6 years; thus, excessive surgery should to be avoided. At present, the patient's family members do not wish to undergo further surgery.

JXG on the upper lip is extremely rare, and to the best of our knowledge, only three previous cases have been

reported in the plastic surgery literature in English.[8–10] In all these cases, the masses were 6–10 mm in diameter and tissue biopsies were performed. Even if JXG is suspected from the clinical presentation, as in the present case, a policy of watchful waiting alone may cause major anxiety to family members if the mass grows rapidly over a short period of time. One option may be to begin by performing a tissue biopsy and obtain a definitive diagnosis, after which the patient can be monitored in anticipation of spontaneous regression. This is because, in most cases, JXG regresses spontaneously and excessive resection should be avoided in surgical treatment. Further investigations of the time required for spontaneous regression involving the observation of a large number of cases for an appropriate monitoring period are required.

CONCLUSIONS

We treated a patient with JXG on the upper lip. JXG on the upper lip is extremely rare, and to the best of our knowledge, this is only the fourth reported case. In most cases, JXG regresses spontaneously and excessive surgery should be avoided. The possibility of JXG should always be considered for masses that rapidly increase in size in infants and young children, and it is important to reach a definitive diagnosis by tissue biopsy.

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